Spinal cord compression associated with pseudohypoparathyroidism

S M Alam MB BS MRCP **W Kelly** MD MRCP
Department of Endocrinology and Diabetes, Middlesbrough General Hospital, Ayresome Green Lane, Middlesbrough TS5 5AZ

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Pseudohypoparathyroidism was described by Albright in 1942[1]. The features include obesity, mental retardation, epilepsy, and hypocalcaemia. Resistance to parathyroid hormone (PTH) was suggested[1], and later verified by PTH infusions. We report a rare neurological association.

Case report

The weight, height and bone age of the patient were above average at 3½ years. At 11 years the patient had epilepsy, controlled by phenytoin. At 23 years he had four weeks of progressive leg weakness, then incontinence of urine and faeces. His appearance suggested Cushing's syndrome, with facial roundness, obesity, weakness, and abdominal striae (Figure 1). He was hypogonadal with undescended testes. He had a spastic paraparesis without a sensory level.

Serum calcium was low, and alkaline phosphatase was raised (Table 1). Plasma bioactive PTH was low, and when 100 ng/l of PTH standard was added to plasma samples from the patient, only 35% was recovered (normal, 50-90%), suggesting an inhibitor to the action of PTH[2]. Bovine PTH intravenously showed impaired phosphate excretion and a sub-normal rise in urinary cAMP. With alpha calcidol therapy, serum calcium became normal. The myelogram showed compression at T1/2, T10/11 and L4/5. Bone histology showed no features of osteomalacia or hyperparathyroidism.

The spinal cord was decompressed by three separate dorsal laminectomies. Very hard bone was noted, but no evidence of extradural or intradural space-occupying lesions. The cord looked normal. There was initially rapid improvement in bladder and bowel function, and a gradual improvement in the function of the legs. With physiotherapy the patient could walk with a frame. He then deteriorated, with weaker legs. Further neurosurgical decompression was attempted, but the spinal cord had infarcted below T10, and he is again paraplegic.

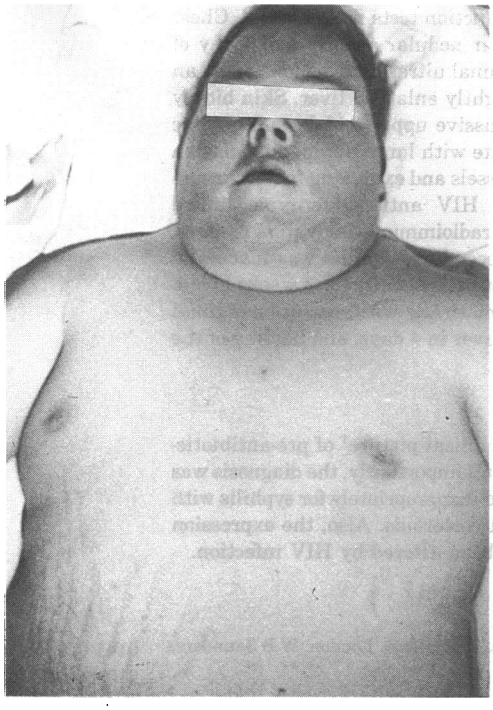

Figure 1. Patient aged 23 years, showing obesity (141 kg) and striae. This classical appearance of pseudohypoparathyroidism resembles Cushing's syndrome, but cortisol levels were not elevated

Table 1. Investigations in a patient with pseudohypoparathyroidism

	Inpatient	*Normal range*
Serum calcium (mmol/l)	1.38	2.1-2.6
Serum albumin (g/l)	45	35-50
Serum inorganic phosphate (mmol/l)	1.75	0.8-1.4
Alkaline phosphatase (u/l)	142	35-115
Plasma immunoactive PTH levels (mean=0.6, SD=0.3, n=6) (u/l)	between 0.3 & 0.9	0.3-1.4
Plasma bioactive PTH concentration (1 ng/l=2.3×10^{-3} IU/l) (ng/l)	0.15	1-15
Urinary calcium excretion (mmol/24 h)	0.34-0.83	2.5-7.5
Phosphate excretion (mmol/24 h)	29	16-50
Serum 25-hydroxyvitamin D (pre-treatment), September 1986 (nmol/l)	7	25-75
Testosterone (nmol/l)	3.1	11-36
Urea and electrolytes	normal	
Urinary free cortisol (nmol/24 h)	220	<300
Plasma cortisol 0900 h (nmol/l)	140	150-600
Free thyroxine index (units)	104	55-180

Discussion

Our patient presented with the typical features of pseudohypoparathyroidism type 1[1]. Treatment with alpha calcidol increased serum calcium, and decreased alkaline phosphatase.

Patients with pseudohypoparathyroidism have been reported to have a spectrum of radiological abnormalities from osteoporosis to osteitis fibrosa cystica and osteomalacia[3]. Sometimes the radiological appearance is normal, but osteomalacia has been suspected on biochemical evidence, and confirmed by bone histology[4]. The normal bone histology and the apparently normal or low immunoactive PTH in our patient may be due to the presence of the PTH inhibitor[2]. There have only been four previous reports of patients with spinal cord compression and pseudohypoparathyroidism[5-8]. These authors reported excessive bone formation in the vertebral canal at C3/4[7] and spondylogenic myelopathy leading to paraplegia[6].

The aetiology of cord compression in pseudohypoparathyroidism is disputed. Calcification of the longitudinal ligaments of the spine may compress the cord[8]. The cord compression may be related to the effects of PTH and vitamin D on the vertebrae and ligaments. It is interesting that X-linked hypophosphataemic rickets may also be complicated by spinal cord compression and extremely hard bone[9], both of which occur in pseudohypoparathyroidism, as in our case[5]. Although he presented with low 25-OH-vitamin D, our patient had no radiological or histological evidence of osteomalacia.

It has also been suggested that the cord compression develops as a result of vitamin D therapy[5]; this seems unlikely, since patients have developed the complication before vitamin D has been given, and myelopathy has not worsened after vitamin D therapy has been commenced. Possibly the bony abnormalities of the spine are genetically determined. The response to surgical decompression of the cord has been disappointing. Therefore, clinicians who follow up these patients should be aware of this complication, and early diagnosis is essential.

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0141-0768/90/010050-02/$02.00/0

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Primary intraocular lymphoma with lymphomatous meningitis

P G Hykin MBBS SHO **J E MacSweeney** MRCP **D Gartry** FRCS **E Graham** MRCP DO *Medical Eye Unit, St Thomas' Hospital, London SE1 7EH*

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We report a patient with ocular lymphoma who developed bilateral seventh nerve palsies due to lymphomatous meningitis without intracerebral involvement. The palsies markedly improved after cerebral irradiation and intrathecal methotrexate. Further studies are required to determine whether repeated cerebrospinal fluid examination and prophylactic cranial and spinal radiotherapy are indicated in patients who present with the disease localized to the eye.

Case report

A 62-year-old man had blurred vision for 2 years, diagnosed as bilateral posterior uveitis. Aetiological investigations were unrewarding. Despite repeated courses of high dose oral prednisolone, 20 months later visual acuity and colour vision deteriorated and he suddenly developed bilateral seventh nerve palsies.

At presentation, right visual acuity was 6/9, left 6/24. No Ishihara colour plates were seen with either eye. Pupillary responses were normal and there was no relative afferent pupillary defect. Both visual fields were markedly constricted. The anterior chambers were quiet. There were numerous large cells in both vitreous chambers, with a resultant hazy fundal appearance. Flourescein angiography showed characteristic pigment epithelial defects. There were marked bilateral seventh nerve palsies (Figure 1) with left hyperacusis. There was no clinical evidence of lymphoma elsewhere.

Cytocentrifuge and immunochemical staining of cerebrospinal fluid revealed β-cell lymphoma cells (Figure 2). Chest X-ray, bone marrow trephine, and computerized axial tomography of the head and abdomen were normal.

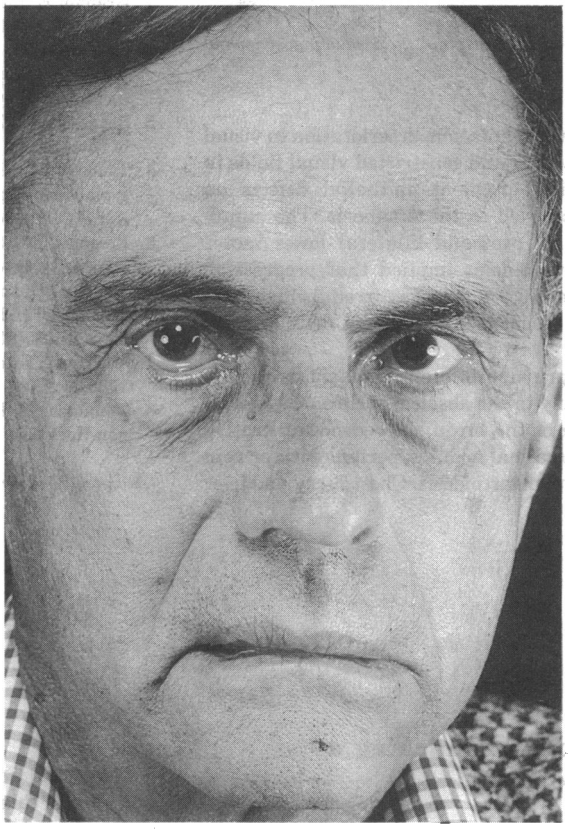

Figure 1. Bilateral seventh nerve palsies

Orbital irradiation (600 cGy) and total cranial irradiation (3750 cGy) were given. After therapy, right visual acuity was 6/6, left 6/18. Both vitreous chambers had cleared. Both facial palsies were markedly improved and the patient remained well for 2 years before progressive dementia and death from subsequent cerebral lymphoma.

Discussion

This patient presented with posterior uveitis of obscure aetiology. Bilateral uveitis, failure to respond to steroids and the high incidence of ocular lymphoma in this age group were suggestive of the diagnosis[1].

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